

H. T. G. from A. W. B.

CASE OF CHRONIC HYDROCEPHALUS

CONNECTED WITH

CANCER AT THE BASE OF BRAIN.

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WITH

AN ACCOUNT OF THE MORBID APPEARANCES.

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THE subject of this case, aged (at the time of his death) eleven years, was admitted into the Royal Infirmary, upon the recommendation of my friend Dr Foulis, on the 26th September 1855, and died on the 12th of November. The following history, the chief features of which I obtained from the mother of the patient, shortly after his admission into the hospital, but many of the more minute particulars from various sources since his decease, I beg, in connection with the very accurate account of the remarkable appearances found upon dissection by Dr Haldane, to offer to the Society, in the belief that the record of a case, in many points of view so instructive, is likely to interest its members.

J. N. was born in the parish of Duddingstone. His father is an agricultural servant, now aged 50, his mother aged 45, both healthy persons, and belonging to families, some of the members of which have attained a remarkable longevity, and have apparently been free from any hereditary disease. The original number of the immediate family was *seven*, and of this *five* survive, at the ages of 18, 16, 13, 8, and 5, all ruddy, healthy looking boys and girls. The only other death which has occurred being that of a sister, aged five years—and now seventeen years ago—from convulsive fits, which were supposed to result from a severe blow sustained upon the head some time previously. At the birth of J. N., two peculiarities were

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noticed, *one*, it may be presumed, of no interest or importance in connection with the case, a ranula, which, within a few days, was appropriately treated and discussed; *the other* of very different signification and import, namely, the *left* eye being very perceptibly smaller than the right, while the latter was of the ordinary or natural size; besides being smaller, it appeared more deeply set in the orbital socket, and very frequent lachrymation gave evidence of its being weaker. In a note, with which I have been favoured by Mr Hill of Portobello, who carefully watched the progress of the case throughout, he says, "the expression and appearance of the left eye was always different from the right, and in early infancy its total want of vision was obvious." It is probable that want of sight in the left eye was congenital, though the determination of its absence was not settled before the age of three, when Mr S., upon whose farm the father of the patient was employed, ascertained that he could not distinguish a watch and certain other objects with which he was familiar, when the right eye was closed. At birth nothing peculiar was noticed in regard to the size of the head; and with the exception of the frontal region being thought broad, nothing abnormal in its conformation. The mother is satisfied that the fontanelles were not longer in closing than in the heads of her other children. Between the ages of three and four, and after the blindness of the left eye had been determined, the following remarkable changes, which I shall first notice in the words of Mr Hill, occurred:—"Both eyes became more prominent, and the left one projected in an extraordinary degree, so as to become at one time completely pushed out of its socket, the eyelids constricting it behind, and giving rise to the most extreme suffering. With some difficulty I was enabled to place it within the eyelids again, and shortly after a large quantity of pus was discharged, which had been collecting behind the eyeball, and the eye gradually assumed the appearance it afterwards retained." The discharge appears, from the testimony of his parents, to have continued during about eight days. About the same time he began to complain of pain, always referred to the back of the head, and a perceptible increase in the size of the cranium occurred. At five years of age, he had measles severely, being the only febrile ailment he ever suffered from, with the exception of a very mild attack of scarlatina, about twelve months before his death. The sight of the right eye, which, in his fifth year, had been becoming gradually impaired, was, in his sixth, totally lost. During these years, besides the kind care of Mr Hill, the little patient was seen by several eminent medical men in Edinburgh, who concurred in believing, that the remarkable appearance presented by the left eye, and the want of vision in both eyes, depended on the existence of a tumour within the cranium. All further concurred in a most gloomy prognosis. In his seventh year, with his head gradually enlarging, and now manifestly abnormally large, he enjoyed good health,—his bodily frame developed proportionally more than in his

earlier years,—he played about, joined heartily in the games of other boys,—was seldom noticed to be taciturn,—and with the exception of the not unfrequent attacks of severe pain in the back part of the head, had no complaint. These headaches were noticed by his mother to be very sudden in their occurrence,—he would join his companions at play, and unexpectedly return to the house, complaining of severe pain always in the same locality, when, after rest for a short period in the horizontal posture, he would rise and again join in play or amusement. At a little more than seven years of age, his health was so good, that, acting under the advice of some kind friends, his parents entered him as a boarder at the “School for Blind Children” in Gayfield Square. Here he continued to reside till very nearly the time when he came under my care in the Infirmary last September. At the Blind School he was frequently seen by Dr Foulis, who very prudently enjoined upon the teachers the propriety of not forcing him on too speedily in his instructions, and particularly cautioned all with whom he there came in contact, to avoid all possible injury to his then too evidently abnormally enlarged cranium.

Upon making inquiry at the Blind School, the teacher, Mr Haig, very kindly favoured me with the following particulars regarding the progress J. N. made while there. He entered the Institution in September 1852, when the teachers and servants of the school were much struck by the appearance of his head. He occasionally complained of pain; but during the three years he spent there, his health was as good as that of any of the other children. He took the same amount of exercise. He was rather selfish and stubborn in his disposition. His education was begun in the school, from the alphabet. It very soon became evident that he was a boy of good abilities,—decidedly above the average of the boys who have been scholars there. He made rapid progress; and agreeably to the advice of Dr Foulis, he was kept back, rather than encouraged, as would have been the case with another boy. For reasons which will appear, when Dr Haldane reads his account of the dissection, I made very special inquiry regarding the integrity of the other senses, as well from Mr Hill, as from the parents of the boy, and his teacher in the Blind School. All agreed in stating that hearing was perfect; but Mr Haig felt quite certain that, for some time past, his sense of smell had become materially impaired. This was opposed to the belief of Mr Hill and the boy’s parents; but it must be remembered that, during the last three years of his life, the teacher in the Blind School had a better opportunity of forming an opinion upon this point than even his parents, from whom, with the exception of a few week’s holidays, he was absent during the whole of that period. Moreover, the illustration which Mr Haig gave me of the manner in which he determined the impairment of this sense, is very striking. It has frequently happened, he informed me, that when the children have been taking walking exercise, they have, as

the blind are, of course, prone to do, placed their feet in dirt, or otherwise come in contact with offensive matter, and, on many occasions, J. N. has done so; while any of the other children who might have done this would have made the discovery for themselves, and remedied it accordingly, this boy never did so; and even after the circumstance was mentioned to him, it was difficult to impress it upon him, owing to the deficiency of smell. Occurrences of this kind, and of a similar nature, led Mr Haig to regard his sense of smell as decidedly impaired. In opposition to this view, it is right, however, to mention, that both the boy's parents, within a few months of his death, saw him smelling an apple, at least holding it to his nose, while they heard him speak of its odour being pleasant. The few weeks of vacation, during last autumn, J. N. spent with his parents; during the earlier part of it, they thought him looking well, though quite certain that the size of the head had very materially increased. Both parents had always regarded him as the cleverest child they had, and, on his return from the Blind School on this occasion, they were much gratified to find that his education had been very considerably advanced. He could now read the Bible with its raised character for the blind, while in knowledge of arithmetic and geography, he had made very considerable advancement. During the month of August, he played about with his companions, being overjoyed to find his way round about his father's cottage and the farm offices in its vicinity,—an occupation in which the blind boy was without a rival. In September, the headaches, from which he had never been entirely exempt, occurred with greater frequency and severity; and being rendered additionally anxious regarding him, on account of a strange drowsiness which frequently overcame him, his parents requested me to admit him into the Infirmary.

The following was his condition at that time. The great enlargement of the head was at once noticed, and the measurements were found to be—*over the vertex, from ear to ear, 1 foot 2 inches; circumference of head, 1 foot 11½ inches.* The left eye was very prominent, but had a dull vacant expression, indicating the absence of sight,—so also the right, which was not prominent. Both pupils were large, but their size did not vary. Pressure over the prominent eye, even to a slight extent, caused much suffering; over the right eye, pressure could be borne. Hearing acute, no complaint of want of smell, nor any evidence of the failure of that sense existed. It was not, however, tested in the way that I now wish it had been done. Face full and somewhat florid. Head plentifully covered with a peculiar woolly looking, sandy-coloured hair. No imperfection in the osseous development of the cranium could be detected; on the contrary, everywhere the bones felt of the usual hardness and firmness. Frontal region broad and prominent. Articulation distinct, and voice loud in replying to questions. Body well covered; fingers and hands smaller than the size and age of

the boy would have indicated. Perfect freedom in locomotion. Only complaint that of severe pain in the occipital region. Sleeps well, occasionally moaning. Appetite good. Bowels regular. Dejections and urine healthy. Respiration and circulation free from any evidence of disease. A few days after admission, the hair of head was removed, and a blister applied to the back of the neck, with some relief to the pain; but owing to its return, and on account of a degree of somnolency, accompanied by slowness of the pulse, he was cupped about the middle of September, and another blister applied, while he took small doses of calomel, until salivation was produced. This resulted in a severer form than was intended, and he suffered a good deal from the irritation it produced. Unquestionably, a considerable degree of relief from the headache followed this treatment, and no farther measures of importance were adopted, save keeping up, in a very mild degree, the action of the mercury, and the renewed application of blisters to the neck. The remarkable intelligence of the boy was a matter of daily observation in the ward, and he very speedily got reconciled to his hospital life, and became a particular favourite with the other patients. The special sense of touch was in him finely developed, as it generally is in the blind, and he frequently astonished gentlemen in the ward by the readiness with which he distinguished the different sizes of some keys which I carried in my pocket, and gave to him for that purpose.

The action of the bowels was always carefully attended to, and laxative enemata were frequently administered. He had never any sickness or vomiting. Occasionally he complained of giddiness as well as of pain in the head. For a week previous to the fatal event, he had been more than usually cheerful and happy, had expressed a desire to walk about the ward, and one day mentioned that his parents, who believed him to be now greatly better, wished him to return home.

On Sunday, the 11th of November, he was, to all appearance, in the same state as during the previous week. I saw him that day, and had a little conversation with him. In the early part of Sunday night he was noticed to be restless; but as his nights had formerly been disturbed, less attention was paid to that circumstance than might otherwise have been the case. Above five A.M. on Monday, the 12th November, he was seized with a violent convulsive fit, and became quite insensible. During two hours he had a succession of fits, then he became quiet, though never sensible. After a recurrence of another convulsion of great severity, about ten A.M., he expired. He was seen, before death, by my friend and house physician, Dr Moore.

Before the Society is put in possession of the very remarkable appearances found upon dissection by Dr Haldane, I would beg, very shortly, to direct attention to the few following points which appear to me of interest in the history of the case:—

1st, The evidence of *congenital* disease within the cranium, as marked by the peculiar aspect of the left eye, and by the want of sight in it, which, though not positively determined till the boy was in his third year, in all probability was absent from birth.

2d, The enlargement of the head, and distension of the cranial bones, was first noticed at the same time as the changes took place in the eyes, *i.e.*, between the third and fourth years, and very evidently point to the intimate connection existing between the tumour and the fluid, which thereafter continued to accumulate. "Chronic hydrocephalus," writes Dr West, "is a morbid condition met with in children at various ages, and coming on under a great variety of circumstances. Sometimes it is congenital, and is then often, though by no means invariably, associated with malformation of the brain. In subsequent childhood, an excess of blood in the brain, or its deficiency, or the existence of some impediment to the circulation through the organ ~~and~~ conditions, all of which have been found to give rise to the effusion of fluid into the cavities of the brain, or upon its surface." Admitting, as it indeed appears necessary to do, with Rokitansky and Vrolik, and with Dr West, that chronic hydrocephalus is not, in many instances, a mere passive dropsy, but that it may be the result of a slow kind of inflammation of the arachnoid, especially of that lining the ventricles; in the case now detailed, it is, I think, very evident that the effusion into the ventricle arose from the impediment to the venous circulation caused by a tumour, the nature and exact relations of which will be described by Dr Haldane. It is rare, says the lamented Dr Valleix, that chronic hydrocephalus, coming on a certain time after birth, is not found in connection with an organic lesion, which accounts for the collection of serum. Such are, for the most part, tumours of different kinds, cancerous, tubercular, cystic. Dr Robert Whytt, a former distinguished professor of medicine in the University of Edinburgh, seems to have been the first physician who directed attention to the possibility of dropsies of the brain being produced by the pressure exerted on the circulation from the presence of tumours. In his "Observations on the Dropsy in the Brain," published in 1768, the following passage occurs:—"A scirrhus tumour of the glandula pituitaria, or in any part contiguous to the ventricles of the brain, by compressing the neighbouring trunks of the absorbent veins, will prevent the due absorption of that fluid which the small arteries constantly exhale, and occasion a dropsy in the brain; in like manner, as a scirrhus liver, spleen, or pancreas, are often the cause of an ascites. As a proof of this, we may observe that M. Petit often found the glandula pituitaria scirrhus in those who died of a dropsy in the ventricles of the brain. In one case I met with, a hard tumour within the right "thalamus nervorum opticorum."

One of the most frequent causes of chronic hydrocephalus, according to M.M. Barthez and Rilliet, is the development of a tumour

within the cranium, ordinarily tubercular, but sometimes also cancerous, or of some other nature. Of chronic hydrocephalus so produced, both these authors and M. Legendre have recorded instances.

3d, and lastly. It is important to note the great intelligence possessed by this boy, the full interest of which will, however, be better appreciated, when the details of the *post-mortem* appearances have apprized the Society of the amount of destruction done to the cerebral substance.

Sectio Cadaveris fifty-one hours after death.

External appearances.—No commencement of putrefaction. Head enlarged; forehead prominent. The measurements corresponded to those taken during life. The left eye was prominent, but otherwise appeared natural. The neck was a little fuller than usual.

Head.—When the scalp was removed, the ossification of the bones of the cranium was found perfect. - The anterior fontanelle was quite filled up; there was a marked depression in that situation. The sutures (particularly the coronal and sagittal) were more marked than natural, but there were no ossa triquetra. The skull cap was very thin, this was particularly the case in regard to the frontal, the anterior part of the parietal, and the squamous portions of the temporal bones. The bone was generally diaphanous, and in places was scarcely more than a line in thickness. The dura-mater having been removed, the surface of the brain was found to be drier and paler than natural. The convolutions were much flattened out, and the intervening sulci were very shallow; this appearance was most distinct on the left side. There was an evident difference in the size of the cerebral hemispheres, the left being markedly the larger. Over the left hemisphere fluctuation was distinct, the right felt soft but solid. On slicing the brain, the left lateral ventricle was very soon reached, its upper wall being under half an inch in thickness; when opened into, a golden yellow coloured serum began to escape. The upper layers of serum were quite clear, the lower were dark and bloody. There was considerable difficulty in removing the fluid from the deeper portions of the ventricle, as the orifice of the pipette became obstructed by what appeared to be a membranous substance. The whole amount of serum in this ventricle was about sixteen ounces. On fully laying open the ventricle, it was found to be very much dilated; its anterior and internal part was occupied by a yellowish red, soft mass, on the surface of which lay several small loose undecolorised clots of blood. This mass, which was partially enclosed in a loose but tolerably strong membrane, appeared to arise from the floor of the ventricle; it was bounded, posteriorly, by the corpus striatum, but extended along the inner margin of this body, and went so far back as to press upon the optic thalamus. As the tumor bulged inwards in the direction of the right hemisphere, the anterior portion of the third ventricle, the septum lucidum, as well as what could be seen of the longitudinal fissure,

were pushed over to the right side. The left corpus striatum, and the optic thalamus, though pressed upon by the tumor, were not involved in it. The whole lining membrane of this ventricle was thicker and tougher than natural.

On examining the right lateral ventricle, its anterior cornu was found very small, evidently from the pressure of the tumor; the posterior cornu was dilated, and, along with the middle cornu, contained about two ounces of serum, chiefly clear, but mixed with a little blood in its lower layers. The parts contained in this ventricle were quite natural. The foramen of Monro was about the size of the tip of the little finger, but appeared partially closed by a portion of the investing membrane of the tumor. The choroid plexus, in either ventricle, was natural. The third ventricle was scarcely enlarged.

The brain having been removed, a mass was found to project from the anterior part of the base, chiefly on the left side. This mass was of a yellowish red colour, and presented a nodulated appearance, some of the nodules being of the size of filberts or small walnuts, but projecting little above the general surface of the tumor. The tumor was accurately bounded, externally and posteriorly, by the fissure of Sylvius of the left side, towards the mesial line it extended as far back as to the corpora albicantia. Anteriorly it reached nearly to the extremity of the anterior lobe. It had not involved the anterior part of the right hemisphere, but had extended towards it, so that the longitudinal fissure was pushed over fully an inch to the right. Farther back, however, the tumor had involved the angular portion of the anterior lobe, situated between the fissure of Sylvius and the longitudinal fissure. The anterior part of the left lateral ventricle had pushed aside the right hemisphere, and so appeared to constitute a part of the base of that division of the brain. Small clots of blood were found along the margin of some parts of the tumor, particularly in the left fissure of Sylvius. Connected with the posterior edge of the mass, about an inch from the commencement of the left fissure of Sylvius, was a small tumor attached to it by a membranous pedicle. It had the appearance of a cyst growing from the investing membrane of the tumor. It was of the size of a large filbert, was of a reddish brown colour mottled with yellow, and had a close resemblance to the polished surface of some agates. Several small blood vessels could be seen to run along its surface.

The mass of the large tumor, as it appeared at the base of the brain, had a firm, almost gristly feeling, the lobules, however, felt soft, almost fluctuating.

The crus cerebri of the left side appeared longer than the other, and the pons was twisted upon itself, being pulled down on the left side towards the cerebellum.

On examining the base of the brain, the olfactory nerves, the left optic nerve, and the optic commissure, could not be made out, they

appeared to have been involved in the tumor. The right optic nerve where it lay upon the crus cerebri was natural.

All the other cranial nerves were uninvolved.

On cutting through the tumor it was found to extend from the base of the brain upwards into the left lateral ventricle. Its lower surface was firm, and of a greyish pink colour, but the surface which appeared in the ventricle, as well as the nodulated portion, was soft. On gently squeezing the latter portions, a milky juice escaped. The cystic tumor connected with its margin contained about two drachms of thin bloody serum. The cerebral matter immediately surrounding the tumor appeared natural. The pituitary gland was flattened out, and appeared a little enlarged, but its structure was healthy.

The dura-mater lining the base of the skull, as well as the bones themselves, were healthy. The left orbital plate of the frontal bone was not thicker than card board.

The left eye was taken out and examined; the opening of the optic nerve into it was very small, the nerve itself was almost entirely atrophied. The cellular tissue behind and around the eye-ball was condensed and firm. The structure of the eye, lens, vitreous humour, etc., seemed natural.

The *neck* was not examined, but the thyroid gland was evidently a little larger than natural.

On opening the *thorax*, the thymus gland was found to occupy the anterior mediastinum; its inferior extremity rested upon the right auricle of the heart. It consisted of two lateral halves (the left being a little the larger) connected together by loose cellular tissue. It was softish, and presented the normal appearance of the unatrophied gland. Its greatest length was $3\frac{3}{4}$ inches, breadth $1\frac{1}{4}$ inches, its weight was 227 grains. The heart was quite healthy. The lungs were normal; they were not congested, their weight was 15 ounces. The bronchial glands were natural. The liver, spleen, and kidneys were quite healthy. The supra renal capsules were no larger than natural. The mesenteric glands were a little enlarged, but contained no abnormal deposit. The intestines were perfectly healthy, the follicular and agminated glands were distinct, but not enlarged. The bladder, prostate and testicles, were natural.

Microscopic Examination.—On examining a drop of the milky juice squeezed from the tumor, it was found to consist chiefly of round and oval nuclei, either loose, or imbedded in a very soft transparent substance. These bodies were about $\frac{1}{2000}$ of an inch in diameter, and contained one or two nucleoli and a little granular matter. In addition to these, though much less numerous, were cells of a round or oval form. These were from $\frac{1}{1500}$ to $\frac{1}{1000}$ of an inch in diameter, and each contained a nucleus exactly similar to those floating about. On the addition of dilute acetic acid, the cell walls became a little paler and more transparent; no change was produced upon the nuclei. The denser portions of the tumor con-

tained much fibrous tissue, combined with a smaller proportion of the cellular elements. The tumor was found to be abundantly supplied with blood vessels, arranged in some places in loops and tufts.

On examining the fluid contained in the ventricles, only blood corpuscles could be detected.

On examination of the thymus gland, abundance of the ordinary corpuscles were seen, with a considerable intermixture of fat.

Remarks.—From the external appearances, as well as from the microscopical structure of this tumor, there can be no doubt as to its cancerous nature. It consisted, however, of two portions, one of which was much harder and firmer than the other. It is, I think, very possible, that the growth was originally of a non-malignant nature, a circumstance which would account for the slow progress it made during the first years of life. This tumor was pretty accurately circumscribed; it was generally perfectly limited, and though it had involved a portion of the other hemisphere, this was rather due to simple extension than to cancerous infiltration. As was formerly mentioned, it presented no tendency to involve the dura-mater or the cranial bones. It is also worthy of remark, that the tumor in the brain was the only local manifestation of the cancerous diathesis. According to Dr Walshe, the cases in which cancer in the brain is unassociated with the same disease in other organs, are about equal in number to those in which it is so associated. Judging from the history of the case, there can be little doubt but that the disease was congenital; it probably commenced in the internal and posterior part of the anterior lobe of the left hemisphere, consequently in the immediate neighbourhood of the left optic nerve. Hence the blindness which appears to have existed from birth. For a long time the tumor increased very slowly, a circumstance explained by the structure of its lower part being firm, and almost cartilaginous. The right optic nerve did not become affected for a period of at least five years. Previously to this, however, the head had been gradually enlarging. The hydrocephalus, which produced this enlargement, was evidently secondary, and was due either to obstruction to return of the blood, produced by the pressure of the tumor, or depended upon a gradually increasing amount of fluid, the result of repeated congestions. The complete ossification of the skull proves that the observation of the boy's parents was correct.

The most doubtful point connected with the case is as to the condition of the sense of smell. From the situation of the tumor, the left olfactory nerve must, I believe, have been affected from birth, the right, in all probability, not for some years later. Reasoning from the *post-mortem* appearances, smell must for some time previous to death have been utterly impossible. And it must be remarked here, that the loss of smell might very readily have escaped the attention of the patient's friends; for, granting that the special sense

was destroyed, common sensation of the mucous membrane of the nostrils unquestionably remained. The circumstance of the boy taking pleasure in holding an apple to his nose, may have been the result of agreeable associations.

Judging from the history, the tumor must have begun to enlarge more rapidly within the last few months; this is explained by the soft structure of the recent growth. The immediate cause of death was, I apprehend, hæmorrhage from one or more of the blood-vessels, with which the growth was abundantly supplied; the appearance of the blood in the ventricle and elsewhere, indicated that it had been recently extravasated, while the occurrence of convulsions fixes the exact date, it being well known that, in the case of children, hæmorrhage into the substance, or upon the surface of the brain, far more frequently gives rise to convulsions than to paralysis.

The unimpaired condition of the boy's mind is highly interesting, as illustrative of the great amount of lesion of the deeper portions of the brain, which is compatible with perfect integrity of the intellectual faculties. The mere flattening out of the convolutions, appears not to prevent their grey matter from performing its functions.

In the account of the *post-mortem* appearances, I have alluded particularly to the condition of the thymus gland, as it has been noticed that in cases of sub-acute and chronic hydrocephalus, this body does not undergo its normal involution. In this case the thymus was considerably larger than is usual at the age of eleven years, and its fatty transformation had made comparatively little progress.





